abnormalities of orbital hypertelorism, ascites and hydrocele. Meanwhile, the maximum distances of tricuspid valve downward displacement measured by STIC-TUI ranged from 0.46cm to 1.23cm, which were similar with the 2DE measurements. The consistency of 2DE and STIC-TUI analyzed by Bland-Altman method suggested these two methods agreed well.

CONCLUSIONS Fetal echocardiography can provide reliable evidence for the diagnosis of Ebstein's anomaly. The maximum distance between the septal hingepoints of tricuspid and mitral valve measured by STIC-TUI was well consistent with the results obtained by 2DE, which indicates STIC-TUI has an important role in the prenatal diagnosis of Ebstein's anomaly.

GW26-e4598

The Results of Cardiac Catheterization and Pulmonary Vasoreactivity Testing in Children with Idiopathic Pulmonary Arterial Hypertension

Yan Zhu, Chen Zhang, Dan Zhang, Wangui Zhao, Hong Gu
Department of Pediatric Cardiology, Beijing Anzhen Hospital, Capital Medical University

OBJECTIVES The aim of this study was to investigate the hemodynamic changes, pulmonary vasodilator agent selection, and responder definition in pediatric patients with IPAH.

METHODS 44 patients with suspected IPAH who admitted to Department of Pediatric Cardiology of Beijing Anzhen Hospital between March 2008 and January 2015 were enrolled. All patients were arranged to receive left and right heart catheterization respectively. The pulmonary vasoreactivity testing with inhalation of pure oxygen and iloprost (PGI_2) were performed during the catheterization. The European Society of Cardiology recommendation criteria (Sitbon criteria) and traditional application criteria (Barst criteria) were used to evaluate the hemodynamic changes.

RESULTS 44 patients with suspected IPAH underwent cardiac catheterization. The mean age was (7.3±4.2) years. 7 patients was excluded from this study with different diagnosis; 7 patients developed pulmonary hypertension crisis during the catheterization. 30 patients received standard cardiac catheterization and pulmonary vasoreactivity testing. The baseline mean pulmonary artery pressure (mPAP) was (66±20) mmHg, and pulmonary vascular resistance index (PVRI) (18.1±7.9) Wood U·m^2. After inhalation of pure oxygen, mPAP decreased to (61±18) mmHg, and PVRI to (18.0±8.5) Wood U·m^2. After inhalation of PGI_2, mPAP decreased to (49±19) mmHg, and PVRI to (12.2±7.3) Wood U·m2 (both $P < 0.001$). According to the Sitbon criteria, the proportion of pure oxygen responders were 6.7% (2/30), while PGI_2 responders were 30% (9/30), and the difference was significant ($P = 0.004$). According to the Barst criteria, the proportion of pure oxygen responders were 16.7% (5/30), while PGI_2, the responders were 50% (15/30), and the difference was significant ($P = 0.001$).

CONCLUSIONS For children with IPAH, cardiac catheterization combined with pulmonary vasoreactivity testing plays an important role in differential diagnosis, severity estimation, and treatment (including the emergency treatment) choices. Pulmonary hypertension crisis is an important complication of cardiac catheterization in pediatric IPAH. Younger age, general anesthesia, crisis history, and poor heart function are important risk factors for pulmonary hypertension crisis. PGI_2 is a relatively ideal agent for vasoreactivity testing in children with IPAH, which has more responders than traditionally used pure oxygen. Results of responders are not completely consistent using different criteria, and comprehensive evaluation should be done according to target of treatment in clinical practice.

GW26-e0237

Prenatal diagnosis of vascular ring by fetal echocardiopraphy combined with STIC technique

Yu Wang, Wei Han, Liu Hong, Haiyan Cao, Yali Yang, Ke Li, Yunfei Hu, Mengmeng Wang, Mingxing Xie
Department of ultrasonography, Union Hospital of Tongji Medical College, Huazhong University of Science and Technology, Hubei Provincial Key Laboratory

OBJECTIVES To evaluate the clinical value of fetal echocardiopraphy combined with STIC for the diagnosis of fetal vascular ring.

METHODS According to the guidelines of ASE, 285 cases of pregnant women were examined by prenatal fetal echocardiography in Union hospital using sequential segmental approach. STIC technique was used to analyze the cardiac volume. All data were collected and stored for further off-line analysis.

RESULTS 1. Among these patients, we found 8 cases of fetus with vascular ring. 5 cases were confirmed by echocardiography after delivery, and other 3 cases were proven by autopsy after induction of labor.

2. 7 out of these 8 cases had the following manifestations in the three vessel and trachea view : aortic arch was on the right side of the trachea while duct artery arch on the left, seen as a "U" shape vascular ring around the trachea.

3. In the three vessel and trachea view, the other 1 case showed the aortic and duct artery arch were both on the left side of the trachea, with the azygos vein flowing into the SVC across the trachea, constituting a "U" shape vascular ring around the trachea.

CONCLUSIONS 1.Fetal vascular ring is a rare congenital heart disease.

2.The three vessel and trachea view is vital for the diagnosis of fetal vascular ring, which consist of dextroaortic arch, left ductus arteriosus and other types of abnormal vessel orientation around the trachea.

3. STIC can show the blood vessel orientation and adjacent structures, which could provide more diagnostic information of fetal vascular ring.

4. Accurate prenatal diagnosis is very important for the prognosis and management of newborns.

GW26-e3538

Pulmonary Hypertension Associated with an Unusual Pattern of Abernethy Malformation - A case reprot and literature review

Xiaolin Zhang, Fangyun Wang
Beijing Children's Hospital

OBJECTIVES We present a case of a 10 month female who was diagnosed with Abernethy Malformation(AM) complicated with Pulmonary Hypertension(PH), whose portal venous blood return has not been described previously. The aim is not only further the knowledgement of AM, but also make physicians aware that, in the absence of other disorders, the AM should also be considered as a determinant of PH.

METHODS We put forward the case report primarily. Then we provide a contemporary review of the literature with a total of 21 patients complicated with PH from 30 series of 200 patients, and analyze the clinical date, including basic information, AM subtype, severity of PH, medical treatment an prognosis.

RESULTS The 10 month female was finally diagnosed with AM by CT angiography, which demonstrated the absence of portal vein and inferior vena cava, the spenic vein and superior mesenteric vein joined and drained directly into the hemiazygos vein without passing the liver. The echocardiography certified the complication of arterial septal defect and moderate PH.

CONCLUSIONS Pulmonary Hypertension is an extremely rare complication of AM, which has a substantial impact on survival and requires focused treatment. So we intend to pay more attention to it.

GW26-e0236

Prenatal diagnosis of pulmonary stenosis/atresia by fetal echocardiography and cardiovascular cast

Yu Wang, Wei Han, Liu Hong, Haiyan Cao, Yali Yang, Ke Li, Yunfei Hu, Mengmeng Wang, Mingxing Xie
Department of ultrasonography, Union Hospital of Tongji Medical College, Huazhong University of Science and Technology, Hubei Provincial Key Laboratory

OBJECTIVES Fetal pulmonary stenosis/atresia is a complicated congenital heart disease. This study aims to investigate the diagnosis of fetal pulmonary stenosis/atresia by fetal echocardiography and cardiovascular cast.

METHODS Forty-three out of 432 cases were diagnosed as congenital pulmonary stenosis/atresia by fetal echocardiography, of which 5 cases associated with other complex malformations underwent labor induction. The fetal hearts with the great vessels were made into cardiovascular cast under patients' consent.

RESULTS ① Five cases of fetal pulmonary stenosis/atresia associated with other complex malformations diagnosed by prenatal echocardiography, were all confirmed by fetal cardiovascular cast.

② Abnormalities diagnosed by echocardiography which cannot be displayed in the fetal cardiovascular casts: aortic regurgitation, mitral